

We claim:

1. A method of treating a subject suffering from a lysosomal storage disorder other than
Fabry Disease caused by a deficiency of a specific protein comprising:
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 - (a) producing said protein or an active fragment thereof in an insect cell culture, and
 - (b) administering a therapeutically effective amount of said protein to said subject.
10. 2. The method of claim 1 wherein said lysosomal storage disorder is selected from the group consisting of Pompe Disease, GM1 gangliosidosis, Tay-Sachs disease, GM2 gangliosidosis: AB Variant, Sandhoff Disease, Gaucher Disease, Krabbe Disease, Niemann-Pick Types A-D, Farber Disease, Wolman Disease, Cholesterol Ester Storage Disease, Hurler Syndrome, Scheie Syndrome, Hurler-Scheie, Hunter Syndrome, Sanfilippo A-D, Morquio A-B, Maroteaux-Lamy, Sly Syndrome,
15 Metachromatic Leukodystrophy, Multiple Sulfatase Deficiency, Sialidosis, I-Cell Disease, Pseudo-Hurler Polydystrophy, Mucolipidosis IV, α -Mannosidosis, β -Mannosidosis, Fucosidosis, Aspartylglucosaminuria, Galactosialidosis, Schindler Disease, Cystinosis, Salla Disease, Infantile Sialic Acid Storage Disorder, Batten Disease, Infantile Neuronal Ceroid Lipofuscinosis, and Prosaposin.
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25. 3. The method of claim 1 wherein said protein is selected from the group consisting of acid α -1,4 glucosidase, acid α -1,6 glucosidase, β -galactosidase, β -hexosaminidase A, GM₂ Activator Protein, β -hexosaminidase A, β -hexosaminidase B, glucocerebrosidase, β -glucosidase, galactosylcerebrosidase, acid sphingomyelinase, acid ceramidase, acid lipase, α -L-iduronidase, iduronate sulfatase, α -N-acetylglucosaminidase, acetyl-CoA-glucosaminide acetyltransferase, N-acetylglucosamine-6-sulfatase, galactosamine-6-sulfatase, arylsulfatase B, β -glucuronidase, arylsulfatase A, arylsulfatase C, α -Neuraminidase, UDP GlcNAc:lysosomal-enzyme N-acetylglucosamine-1-phosphotransferase, neuraminidase, α -mannosidase, β -mannosidase, α -L-fucosidase, N-aspartyl- β -glucosaminidase, protective protein/cathepsin A (PPCA), α -N-acetyl-

galactosaminidase, cystine transport protein, sialic acid transport protein, palmitoyl-protein thioesterase, and Saposins A-D.

4. The method of claim 1 wherein said protein is produced in an insect cell culture using
5 a baculovirus expression system.
5. The method of claim 1 wherein said insect cell culture is derived from the species
Spodoptera frugiperda.
- 10 6. The method of claim 5 wherein said insect cell culture is an Sf9 cell culture.
7. A method of treating a subject with a protein other than α -galactosidase that is therapeutically active when present in a macrophage comprising:
15 (a) producing said protein in an insect cell culture; and
(b) administering a therapeutically effective amount of said protein to said subject.
8. A pharmaceutical composition comprising a protein useful for treating a lysosomal storage disorder other than Fabry disease that is selectively imported into
20 macrophages when administered to a subject and a pharmaceutically acceptable carrier, wherein said protein is produced in an insect cell culture.
9. The composition of claim 8 wherein said lysosomal storage disorder is selected from the group consisting of Pompe Disease, GM1 gangliosidosis, Tay-Sachs disease,
25 GM2 gangliosidosis: AB Variant, Sandhoff Disease, Gaucher Disease, Krabbe Disease, Niemann-Pick Types A-D, Farber Disease, Wolman Disease, Cholesterol Ester Storage Disease, Hurler Syndrome, Scheie Syndrome, Hurler-Scheie, Hunter Syndrome, Sanfilippo A-D, Morquio A-B, Maroteaux-Lamy, Sly Syndrome, Metachromatic Leukodystrophy, Multiple Sulfatase Deficiency, Sialidosis, I-Cell
30 Disease, Pseudo-Hurler Polydystrophy, Mucolipidosis IV, α -Mannosidosis, β -Mannosidosis, Fucosidosis, Aspartylglucosaminuria, Galactosialidosis, Schindler

Disease, Cystinosis, Salla Disease, Infantile Sialic Acid Storage Disorder, Batten Disease, Infantile Neuronal Ceroid Lipofuscinosis, and Prosaposin.

10. The composition of claim 8 wherein said protein is selected from the group consisting
5 of acid α -1,4 glucosidase, acid α -1,6 glucosidase, β -galactosidase, β -hexosaminidase A, GM₂ Activator Protein, β -hexosaminidase A, β -hexosaminidase B, glucocerebrosidase, β -glucosidase, galactosylcerebrosidase, acid sphingomyelinase, acid ceramidase, acid lipase, α -L-iduronidase, iduronate sulfatase, α -N-acetylglucosaminidase, acetyl-CoA-glucosaminide acetyltransferase, N-
10 acetylglucosamine-6-sulfatase, galactosamine-6-sulfatase, arylsulfatase B, β -glucuronidase, arylsulfatase A, arylsulfatase C, α -Neuraminidase, UDP GlcNAc:lysosomal-enzyme N-acetylglucosamine-1-phosphotransferase, neuraminidase, α -mannosidase, β -mannosidase, α -L-fucosidase, N-aspartyl- β -glucosaminidase, protective protein/cathepsin A (PPCA), α -N-acetyl-
15 galactosaminidase, cystine transport protein, sialic acid transport protein, palmitoyl-protein thioesterase, and Saposins A-D.
11. The composition of claim 8 wherein said insect cell culture comprises cells derived
from the species selected from the group consisting of *Spodoptera frugiperda* and
20 *Tricoplusia ni*.
12. The composition of claim 11 wherein said cells are *Spodoptera frugiperda* Sf9 cells.
13. The composition of claim 8 wherein said protein is produced in the cell culture using
25 a baculovirus expression system.
14. A method for producing a protein associated with a lysosomal storage disorder other
than α -galactosidase, protective protein/cathepsin A (PPCA), cathepsin B, cathepsin
S, β -galactosidase, β -hexosaminidase B, neuraminidase, lysosomal acid lipase,
30 prorenin, glucocerebrosidase and lysosomal acid alpha-glucosidase in a form that is

selectively imported into macrophages when administered to a subject comprising producing said protein in an insect cell culture.

15. The method of claim 14 wherein said lysosomal storage disorder is selected from the group consisting of GM2 gangliosidosis: AB Variant, Sandhoff Disease, Krabbe Disease, Niemann-Pick Types A-D, Farber Disease, Hurler Syndrome, Scheie Syndrome, Hurler-Scheie, Hunter Syndrome, Sanfilippo A-D, Morquio A, Maroteaux-Lamy, Sly Syndrome, Metachromatic Leukodystrophy, Multiple Sulfatase Deficiency, I-Cell Disease, Pseudo-Hurler Polydystrophy, Mucolipidosis IV, α -Mannosidosis, β - Mannosidosis, Fucosidosis, Aspartylglucosaminuria, Schindler Disease, Cystinosis, Salla Disease, Infantile Sialic Acid Storage Disorder, Batten Disease, Infantile Neuronal Ceroid Lipofuscinosis, and Prosaposin.
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16. The method of claim 14 wherein said protein is selected from the group consisting of GM₂ Activator Protein, β -hexosaminidase A, β -hexosaminidase B, galactosylceramidase, acid sphingomyelinase, acid ceramidase, α -L-iduronidase, iduronate sulfatase, α -N-acetylglucosaminidase, acetyl-CoA-glucosaminide acetyltransferase, N-acetylglucosamine-6-sulfatase, galactosamine-6-sulfatase, arylsulfatase B, β -glucuronidase, arylsulfatase A, arylsulfatase C, UDP GlcNAc:lysosomal-enzyme N-acetylglucosamine-1-phosphotransferase, neuraminidase, α -mannosidase, β -mannosidase, α -L-fucosidase, N-aspartyl- β -glucosaminidase, α -N-acetyl-galactosaminidase, cystine transport protein, sialic acid transport protein, palmitoyl-protein thioesterase, and Saposins A-D,
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25. 17. A protein-conjugate complex that is selectively imported into macrophages when administered to a subject wherein the protein component of said protein-conjugate complex is produced in an insect cell culture.
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18. A method for increasing the ability of a cell to uptake a protein produced in an insect cell culture comprising causing said cell to express a mannose receptor on its membrane.

19. A system for targeting a protein to a desired cell comprising:

- (a) causing said cell to express a mannose receptor on its membrane;
- 5 (b) producing said protein in an insect cell culture; and
- (c) contacting said protein with said cell.

20. In a method for purifying a protein produced in an insect cell culture using a
Concanavalin A-Sepharose column, an improvement comprising the use of a buffer
10 containing methyl- α -D-manno-pyranoside to elute said protein from said
Concanavalin A-Sepharose column.